Objective: To provide a review of the current information on the etiology, clinical presentation, management, and outcome of pilomatrixoma of the head and neck in children.

Design: Retrospective review.

Setting: A tertiary care pediatric center.

Patients: Fifty-one pediatric patients with a diagnosis of pilomatrixoma of the head and neck.

Intervention: All patients underwent excision of pilomatrixoma from January 1997 to March 1999. A total of 55 tumors were studied.

Results: A preponderance of girls (n=36; 71%) presented with this condition. The average age at diagnosis was 5.7 years, and the average size of the lesion was 1 cm. The skin of the cheek and the periorbital area were the most commonly involved sites. Only 27 lesions (49%) had a correct preoperative diagnosis. Two (4%) of 55 tumors recurred after complete surgical excision.

Main Outcome Measures: The age and sex of the patient, preoperative diagnosis, time elapsed before diagnosis, site and size of the tumor, length of follow-up, presence of multiple or previous pilomatrixomas, and recurrence.

Conclusions: Preoperative diagnosis may be improved with increased awareness of pilomatrixoma, a common, benign skin tumor in children. Clinical findings will aid in an accurate diagnosis. Recurrence after complete local excision is rare.


In 1880, Malherbe and Chenantais first described pilomatrixomas, then thought to arise from sebaceous glands and called calcifying epitheliomas of Malherbe. In 1961, Forbis and Helwig proposed the term pilomatrixoma to avoid a connotation of malignancy. At this time, the origin of pilomatrixoma from hair matrix cells was discovered.

Pilomatrixomas commonly occur in children and most frequently in the head and neck region. They are the second most commonly excised superficial masses in children after epidermoid cysts and excluding lymph nodes. However, pilomatrixomas are frequently misdiagnosed and/or missed in the differential diagnosis. Surgical removal is curative. Recurrence after complete local excision is rare. Malignancy has been rarely reported.

The lack of discussion of pilomatrixoma in the otolaryngology literature despite its frequent occurrence prompted this review. The objective of this study is to provide information on the etiology, clinical presentation, management, and outcome of pilomatrixoma of the head and neck in children. We report the results of our own study, in which we analyzed data from 51 children with a diagnosis of pilomatrixoma of the head and neck, a total of 55 tumors; and we provide a review of the literature. The clinical presentation, tumor behavior, treatment, and outcome of pilomatrixoma are reviewed.

RESULTS

There were 51 patients with a total of 55 pilomatrixomas of the head and neck region; 36 (71%) were girls, 15 (29%) were boys (ratio, 2.4:1). Patient age at diagnosis varied from 8 months to 16 years, with 35 patients (69%) presenting with the condition at younger than 7 years; 37 (72%) were diagnosed within 6 months of developing the lesion.

Figure 1 shows the sites of pilomatrixoma occurrence. Seventeen lesions
(31%) occurred around the eye, including the brow and periorbital area, 15 (27%) in the cheek region, and 7 (13%) in the preauricular region.

The varied preoperative diagnoses are depicted in Figure 2. A correct preoperative diagnosis was made 49% of the time. The most common misdiagnosis was a dermoid. Six patients (12%) reported tenderness and 5 (9%) reported a history of trauma in the region of the tumor.

All tumors were slow growing. A bluish hue or telangiectasia was found in 17 lesions (31%); 5 patients (10%) had multiple pilomatrixomas at the time of diagnosis, including areas outside of the head and neck region. Three patients (6%) had a history of a previous pilomatrixoma. All lesions were surgically excised (1 patient underwent attempted removal with liquid nitrogen and experienced recurrence; surgical excision was then performed). Three lesions (6%) recurred overall; after complete surgical excision, 2 lesions (4%) recurred, which is similar to the literature.3

Figure 1. Areas of occurrence of 55 pilomatrixomas.

Figure 2. Preoperative diagnosis of masses (N=55) prior to excision and pathological examination (number of masses/percentage of total pilomatrixoma tumors).

Figure 3. Pilomatrixoma showing shadow cells with naked nuclei and abundant cytoplasm (hematoxylin-eosin, original magnification ×200).

Figure 4. Pilomatrixoma showing inferior and central basaloid cells becoming larger islands of shadow cells (hematoxylin-eosin, original magnification ×100).

Pilomatrixomas are of ectodermal origin and arise from the outer root sheath cell of the hair follicle.3,5 They arise in the lower dermis and form a connective tissue capsule. Shadow or ghost cells, with a central unstained area representing a shadow of a lost nucleus, are common.6 (Figure 3). Basaloid cells have a round or elongated basophilic nucleus and scant cytoplasm at the periphery of epithelial islands6 (Figure 4). Calcium deposition and a foreign body reaction commonly occur,7 and ossification has been reported.6

The diagnosis of pilomatrixoma can be made clinically if the characteristics of the tumor are known (Figure 5): pilomatrixoma lesions slide freely over the underlying area. Graham and Merwin8 described the “tent sign,” elicited by stretching the skin over the pilomatrixoma tumor to feel the irregular surface of the mass. There is no associated lymphadenopathy. A blue discoloration may be seen. We have found the most common locations of pilomatrixoma to be the cheek area and, as Orlando et al7 have noted, the periorbital area. There may be a history of regional trauma prior to developing the tumor, as was found in 5 (9%) of our patients (which matches the results of the literature3). The significance of this is unknown.

The pilomatrixoma tumor commonly occurs in the head and neck region of children. Our age of presentation was similar to that in the literature, with most patients presenting at younger than 7 years.3,7 We found multiple tumors in 5 (10%) of our patients, which is higher

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than the 2% to 3.5% reported in the literature.6 Multiple pilomatrixomas have been associated with Gardner syndrome, Steinert disease, myotonic dystrophy, and sarcoidosis,3-11 although these diseases were not found in our patients. We also found a 71% female preponderance (36 girls vs 15 boys) that has not been found by others.3,7

The differential diagnosis of pilomatrixoma is varied. Pilomatrixoma should be differentiated from epidermal and dermoid cysts. Epidermal cysts are firm, round, and mobile, and they have normal overlying skin. They also present in an older age group, both adolescents and adults.7 Dermoid cysts are firmly attached to underlying tissue and are often found in children. Neither presents with irregular nodules on the skin as pilomatrixoma does.

Diagnostic tests and imaging studies are often unnecessary in the workup of a superficial, benign skin lesion such as pilomatrixoma. However, tests are sometimes done to exclude the diagnosis of malignancy or to determine the depth of a lesion. Pilomatrixoma in the parotid or preauricular region may be a diagnostic dilemma. If the mass is of branchial or parotid origin, careful dissection from the parotid gland may be necessary, and preoperative knowledge of the extent of the lesion may be helpful.

Fine-needle aspiration may reveal the presence of ghost cells, basaloid cells, and/or calcium deposition in the mass, which findings are diagnostic of pilomatrixoma.12 However, without the presence of ghost cells in the aspirate, the diagnosis may be misleading.12 On routine soft tissue radiographs, which have fallen out of use since advent of superior imaging techniques for soft tissue masses, pilomatrixoma may appear as a regularly contoured mass with homogeneous speckles and occasional dense foci of calcification.12 Fink and Berkowitz12 found ultrasound to be particularly helpful in children to determine the relation of the mass to the parotid. This method does not require sedation or anesthesia. Computed tomography and magnetic resonance imaging will add greater detail to the surrounding structures and depth of the lesion, but these imaging techniques are costly and may require sedation or the use of anesthesia in children.

Malignant pilomatrixoma has never been reported in children and was not found in our series. Black et al6 report the clinical behavior of pilomatrix carcinoma in adults to resemble that of basal cell carcinoma in its potential to metastasize. Treatment is wide local excision. Reconstruction may be best deferred for 1 year to allow for close observation for recurrence. The role of radiation is unknown owing to the small number of reported cases, but may help in locoregional control.6

The treatment of choice and standard therapy for benign pilomatrixoma is complete surgical excision. Occasionally, overlying skin will need to be excised secondary to tumor adherence to the dermis. Morales and McGoey14 have advocated incision and curettage for cosmetic preservation in large tumors or for those in exposed areas. No recurrence was found in their series. Our series noted that 2 tumors (4%) recurred after complete surgical excision. One lesion in our study failed to respond to liquid nitrogen therapy and required surgical excision.

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REFERENCES


